
CANCER FACTS

National Cancer Institute • National Institutes of Health
Department of Health and Human Services

Paget's Disease of the Breast: Questions and Answers

1. What is Paget's disease of the breast?

Paget's disease of the breast is an uncommon type of cancer that occurs in 1 to 4 percent of all people with breast cancer. It is sometimes called mammary Paget's disease. Paget's disease of the breast can develop in men, but it is very rare.

This type of cancer was named after Sir James Paget, a scientist who noted an association between changes in the appearance of the nipple and underlying breast cancer. A number of other diseases have also been named after Sir James Paget, including Paget's disease of the bone, which involves genetic changes that increase the risk for osteosarcoma (cancer of the bone). This fact sheet discusses only Paget's disease of the breast. Information about other types of cancer is available from the Cancer Information Service (CIS) at 1-800-4-CANCER (1-800-422-6237).

Scientists do not know exactly how Paget's disease of the breast occurs, but two major theories have been suggested. In one theory, cancer cells called Paget cells break off from a tumor (an abnormal mass of tissue) in the breast and move through the milk ducts in the breast to the surface of the nipple. In the other theory, the skin cells of the nipple spontaneously become cancerous Paget cells.

2. What are the symptoms of Paget's disease of the breast?

Symptoms of Paget's disease of the breast include itching, burning, redness, and scaling of the skin on the nipple and areola. The areola is the circular area of darker-colored skin that surrounds the nipple. There may be a bloody discharge from the nipple, and the nipple may appear flattened against the breast. In up to 30 percent of cases, however, there are no visible skin changes. Almost half of all patients with Paget's disease of the breast also have a lump in the breast that can be felt at the time of diagnosis. It is important to see a health care provider about any of these symptoms, or if the symptoms

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do not completely disappear after treatment. They may be caused by Paget's disease of the breast, other types of breast cancer, or a less serious skin condition.

3. How is Paget's disease of the breast diagnosed?

If the health care provider suspects Paget's disease, a sample of any nipple discharge may be examined under a microscope for Paget cells, or a biopsy of the nipple will be done. In a biopsy, the doctor removes a small sample of nipple tissue. A pathologist examines the tissue under a microscope to see if Paget cells are present.

Most people with Paget's disease of the breast also have an underlying breast cancer. That is why the health care provider usually orders a mammogram (x-ray of the breast). However, women with symptoms of Paget's disease who do not have a lump that can be felt often have normal mammograms. These women may need to have other breast imaging techniques, such as ultrasound or MRI (magnetic resonance imaging). In an ultrasound, high-frequency sound waves that humans cannot hear are bounced off tissues and internal organs. Their echoes produce a picture called a sonogram. In an MRI, a magnet linked to a computer creates detailed pictures of areas inside the breast.

4. What type of breast cancer is associated with Paget's disease of the breast?

People with Paget's disease who **do not** have a breast lump that can be felt usually have a condition called ductal carcinoma *in situ* (DCIS). DCIS is also known as intraductal carcinoma. In DCIS, abnormal cells are present only in the lining of the milk ducts in the breast, and have not invaded surrounding tissue or spread to the lymph nodes. People with Paget's disease of the breast who **do** have a lump that can be felt at the time of diagnosis usually have invasive or infiltrating ductal carcinoma. The cancer has spread to nearby tissue, lymph nodes under the arm, or other parts of the body.

5. How is Paget's disease of the breast treated?

Modified radical mastectomy is the usual treatment for Paget's disease when the patient has an underlying breast cancer, or when the cancer has spread beyond the central portion of the breast behind the nipple. In this operation, the surgeon removes the breast, some of the lymph nodes under the arm, and the lining over the chest muscles. The surgeon may also remove part of the chest wall muscles.

Other treatment options may be available if no underlying cancer is apparent, or when the cancer is located only in the central portion of the breast behind the nipple. Some of these patients receive radiation therapy by itself, without breast-conserving surgery, while other patients have breast-conserving surgery, which may or may not be followed by radiation therapy. Breast-conserving surgery for patients with Paget's disease of the breast involves removing all of the nipple and areola and some of the breast tissue underneath.

For patients undergoing mastectomy, the doctor will perform a biopsy of the lymph nodes. The doctor may not recommend a lymph node biopsy for patients undergoing breast-conserving surgery. Adjuvant treatment (treatment that is given in addition to surgery to prevent the cancer from coming back) may be part of the treatment plan if cancer cells have spread to the lymph nodes. Adjuvant therapy may include chemotherapy, radiation treatment, and/or hormonal treatment.

6. Are clinical trials (research studies) available for people with Paget's disease of the breast?

Yes. Clinical trials are in progress for all types of breast cancer. These studies are designed to find new treatments and better ways to use current treatments. Before any new treatment can be recommended for general use, doctors conduct clinical trials to find out whether the treatment is safe for patients and effective against the disease.

People interested in taking part in a clinical trial should talk with their health care provider. Information about clinical trials is available from the Cancer Information Service (CIS) at 1-800-4-CANCER (1-800-422-6237). Information specialists at the CIS use PDQ®, NCI's cancer information database, to identify and provide detailed information about specific ongoing clinical trials. Patients also have the option of searching for clinical trials on their own. The NCI's Web site, located at <http://cancer.gov/> on the Internet, provides information about clinical trials and links to PDQ.

7. Where can a person find more information about breast cancer and its treatment?

The NCI has several other resources that readers may find helpful, including the following:

- *What You Need To Know About™ Breast Cancer* and *Understanding Breast Cancer Treatment: A Guide for Patients* are booklets that provide information about types of breast cancer, staging, and treatment options;
- *Taking Part in Clinical Trials: What Cancer Patients Need To Know* describes how research studies are carried out and explains their possible benefits and risks; and
- *Questions and Answers About Adjuvant Therapy for Breast Cancer* has more information about adjuvant therapy.

These and other resources can be obtained by calling the CIS, or through the NCI Publications Locator Web site at <http://cancer.gov/publications> on the Internet.

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Sources of National Cancer Institute Information

Cancer Information Service

Toll-free: 1-800-4-CANCER (1-800-422-6237)

TTY (for deaf and hard of hearing callers): 1-800-332-8615

NCI Online

Internet

Use <http://cancer.gov> to reach the NCI's Web site.

LiveHelp

Cancer Information Specialists offer online assistance through the *LiveHelp* link on the NCI's Web site.

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